



## Complete Summary

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### GUIDELINE TITLE

Diagnosis and management of aortic dissection.

### BIBLIOGRAPHIC SOURCE(S)

Erbel R, Alfonso F, Boileau C, Dirsch O, Eber B, Haverich A, Rakowski H, Struyven J, Radegran K, Sechtem U, Taylor J, Zollikofer C, Klein WW, Mulder B, Providencia LA. Diagnosis and management of aortic dissection. Eur Heart J 2001 Sep;22(18):1642-81. [295 references]

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## SCOPE

### DISEASE/CONDITION(S)

Aortic dissection

### GUIDELINE CATEGORY

Diagnosis  
Evaluation  
Management  
Treatment

### CLINICAL SPECIALTY

Cardiology  
Emergency Medicine  
Family Practice  
Internal Medicine  
Surgery

### INTENDED USERS

Physicians

## GUIDELINE OBJECTIVE(S)

To provide recommendations for the diagnosis and management of patients with aortic dissection

## TARGET POPULATION

Patients with aortic dissection

## INTERVENTIONS AND PRACTICES CONSIDERED

### Initial Management

1. Clinical evaluation including assessment of clinical features, differential diagnosis, and physical examination
2. Electrocardiogram (ECG), chest x-ray, and laboratory tests
3. Blood pressure monitoring and control with beta-blockers (e.g., propranolol, esmolol, metoprolol, atenolol, labetalol), calcium antagonists (e.g., verapamil, diltiazem, nifedipine), and vasodilators (e.g., sodium nitroprusside)
4. Pain relief with morphine sulfate
5. Intubation and ventilation for profound haemodynamic instability
6. Transoesophageal echocardiography as initial diagnostic test
7. Pericardiocentesis

### Diagnostic Imaging

1. Computed tomography (CT)
2. Magnetic resonance imaging (MRI)
3. Angiography
4. Evaluation of pressure difference with pulsed Doppler and continuous wave Doppler
5. Analysis of disease extent by echocardiography combined with duplex sonography, abdominal sonography, or intravascular ultrasound
6. Classification of disease via transoesophageal/transthoracic echocardiography, computed tomography, or magnetic resonance imaging
7. Assessment of arch vessel, aortic side branch, or iliac/femoral artery involvement via angiography, duplex sonography, helical computed tomography, magnetic resonance imaging, or transoesophageal echocardiography

### Additional Laboratory Testing

1. C-reactive protein measurement
2. Genetic testing (family studies for fibrillin-1 [FBN-1] gene and other mutation studies)

### Surgical and Interventional Therapy

1. Implantation of composite graft (or alternatively xenografts, allografts, or autografts)
2. Valve sparing operations and/or aortic root remodeling
3. Percutaneous stenting
4. Percutaneous fenestration

#### Follow-up

1. Life-long beta-blockade
2. Periodic imaging studies
3. Prophylactic repair of aortic root
4. Reoperation as necessary

#### MAJOR OUTCOMES CONSIDERED

- Accuracy, specificity and sensitivity of diagnostic imaging techniques
- Complications of surgical and interventional therapies
- Occurrence of aortic rupture and development of pericardial effusion
- Restoration of flow to obstructed vessels
- Mortality and morbidity of aortic dissection

### METHODOLOGY

#### METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

#### DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Not stated

#### NUMBER OF SOURCE DOCUMENTS

Not stated

#### METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

#### RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Levels of Evidence:

- A. Data derived from multiple randomized clinical trials or meta-analyses
- B. Data derived from a single randomized trial or non-randomized studies
- C. Consensus opinion of the experts

#### METHODS USED TO ANALYZE THE EVIDENCE

## Systematic Review

### DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

### METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

### DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

Provisional guidelines were prepared by the European Society of Cardiology (ESC) Task Force on Aortic Dissection, as suggested by the committee for Scientific Clinical Initiatives and approved by the ESC Board at its meeting on 17 June 1997.

This Task Force consists of 11 members, including representatives of the European Association of Radiology, and the European Society of Pediatric Cardiology, as well as one member appointed by the American College of Cardiology (ACC) in order to gain ACC endorsement. The members were all appointed by the Board of the ESC upon suggestions made by the committee for Scientific Clinical Initiatives. In addition, controversial issues were discussed between the members on an e-mail platform as well as via telephone conferences. Review of the literature and position papers were then prepared.

### RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Class I: when there is evidence for and/or general agreement that the procedure or treatment is useful.

Class II: when usefulness of the procedure or treatment is less well established or divergence of opinion exists among the members of the Task Force.

Class III: when the procedure or treatment is not useful and in some cases may be harmful.

### COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

### METHOD OF GUIDELINE VALIDATION

Internal Peer Review

### DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Provisional guidelines were prepared by the European Society of Cardiology Task Force on Aortic Dissection, as suggested by the Committee for Scientific Clinical Initiatives and approved by the European Society of Cardiology Board at its meeting on 17 June 1997. This Task Force consists of 11 members. At the request of the Committee of Scientific Clinical Initiatives, the Task Force delineations were presented at the Congress of the European Society of Cardiology in August 1999 in the conference on emerging guidelines. Finally, the document was distributed for correction and endorsement to all members and intermittently reviewed for consistency by internal reviewers.

## RECOMMENDATIONS

### MAJOR RECOMMENDATIONS

The class of recommendations (I-III) and level of evidence (A-C) are defined at the end of the "Major Recommendations" field.

#### Clinical Management

- Aortic dissection has a wide range of clinical presentations. Clinical suspicion is required for quick risk stratification and management.
- Monitoring of heart rate and blood pressure is indicated. Beta-blocking therapy and blood pressure lowering medications are started, as well as sedation and analgesic therapy.
- Monitoring in the intensive care unit should be initiated. Fluid replacement may be important. Use a separate line for blood pressure lowering. Start with beta-blocking agents and add sodium nitroprusside, if necessary.
- In case of haemodynamic instability, the patient should be intubated and transferred to the operating theatre. Transoesophageal echocardiography may be performed prior to transfer or in the operating theatre.

#### Recommendations for Initial Management of Patients With Suspected Aortic Dissection

##### Class I:

- Detailed medical history and complete physical examination (whenever possible) (Level of Evidence C)
- Intravenous line, blood sample (creatinine kinase [CK], TnT(I), myoglobin, white blood cell, D-dimer, haematocrit, lactate dehydrogenase [LDH]) (Level of Evidence C)
- Electrocardiogram: documentation of ischaemia (Level of Evidence C)
- Heart rate and blood pressure monitoring (Level of Evidence C)
- Pain relief (morphine sulfate) (Level of Evidence C)
- Reduction of systolic blood pressure using beta-blockers (intravenous [IV] propranolol, metoprolol, esmolol or labetalol) (Level of Evidence C)
- Transfer to intensive care unit (Level of Evidence C)
- In patients with severe hypertension additional vasodilator (intravenous sodium nitroprusside to titrate blood pressure to 100 to 120 mmHg) (Level of Evidence C)

Class II:

- In patients with obstructive pulmonary disease, blood pressure lowering with calcium channel blockers (Level of Evidence C)
- Imaging in patients with electrocardiographic signs of ischaemia before thrombolysis if aortic pathology is suspected (Level of Evidence C)

Class III:

- Chest x-ray (Level of Evidence C)

#### Recommendations for Management of Haemodynamically Unstable Patients With Suspected Aortic Dissection

Class I:

- Profound haemodynamic instability: intubation and ventilation (Level of Evidence C)

Class II:

- Transoesophageal echocardiography as the sole diagnostic procedure -- call surgeon (Level of Evidence C)
- Surgery-based on findings of cardiac tamponade by transthoracic echocardiography (Level of Evidence C)

Class III:

- Pericardiocentesis (lowers intrapericardial pressure [recurrent bleeding!]) (Level of Evidence C)

#### Diagnostic Requirements

##### Essential Information in Evolving Acute Aortic Dissection

- Confirmation of diagnosis
- Tear localization
- Extent of aortic dissection
- Classification of aortic dissection (see below)
- Indicators of emergency

##### Additional Information Not Immediately Necessary

- Arch vessel and side branch involvement
- No laboratory tests except for routine parameter assessment

##### Classification of Aortic Dissection

#### Stanford Classification

Type A: dissection of the ascending and descending aorta

Type B: dissection of the descending aorta

#### DeBakey Classification

Type 1: dissection of the entire aorta

Type 2: dissection of the ascending aorta

Type 3: dissection of the descending aorta

#### New Classification

Class 1: classical aortic dissection with an intimal flap between true and false lumen

Class 2: medial disruption with formation of intramural haematoma/haemorrhage

Class 3: discrete/subtle dissection without haematoma, eccentric bulge at tear site

Class 4: plaque rupture leading to aortic ulceration, penetrating aortic atherosclerotic ulcer with surrounding haematoma, usually subadventitial

Class 5: iatrogenic and traumatic dissection

Classes 1 through 5 represent a subdivision to the Stanford or DeBakey classifications

#### Imaging Modalities

##### Transthoracic/Transesophageal Echocardiography (TTE/TEE)

- Transoesophageal echocardiography in addition to transthoracic echocardiography can be used for decision making in the emergency room or even operating theatre in acute aortic dissection with high accuracy. Pitfalls have to be taken into account. The high resolution enables the diagnosis also of intramural haematoma, plaque ulceration, as well as traumatic aortic injury. When more spatial resolution is necessary, computed tomography or magnetic resonance imaging are used in addition.

##### Computed Tomography (CT)

- Computed tomography is the technique used most often in patients with suspected aortic dissection.
- The sensitivity is greater than 90%, specificity greater than 85%. The extent, localization and side branch involvement of aortic dissection can be assessed, signs of emergency detected. Limitations are related to diagnosis of aortic regurgitation, tear localization as well as detection of intimal tears and subtle/discrete aortic dissection (Class 3).

## Magnetic resonance imaging (MRI)

- Magnetic resonance imaging has the highest accuracy and sensitivity as well as specificity (nearly 100%) for detection of all forms of dissection (Classes 1, 2 and 4, 5) except subtle/discrete forms (Class 3). Distribution and availability is limited particularly in emergency situations. Most often magnetic resonance imaging is used in stable haemodynamic conditions and chronic aortic dissection for follow-up. Magnetic resonance imaging provides excellent visualization of tear localization, aortic regurgitation, side branch involvement and complications.

## Aortography

- Aortography is highly valuable to diagnose classical aortic dissection, but limitations are obvious in dissection subtypes such as non-communicating aortic dissection and intramural haematoma and haemorrhage formation (Class 2) as well as plaque rupture (Class 4). Aortography is the standard technique for guiding interventions in aortic dissection.

## Recommendations for Diagnostic Imaging in Acute Aortic Dissection

### Class I:

- Transthoracic echocardiography followed by transoesophageal echocardiography (Level of Evidence C)
- Computed tomography (Level of Evidence C)
- Contrast angiography to define anatomy in visceral malperfusion and to guide percutaneous interventions (Level of Evidence C)

### Class IIa:

- Contrast angiography in stable patients (Level of Evidence C)
- Magnetic resonance imaging (Level of Evidence C)
- Intravascular ultrasound (Level of Evidence C)

### Class IIb:

- Computed tomography if detection of tears is crucial (Level of Evidence C)
- Contrast angiography in haemodynamically unstable patients (Level of Evidence C)
- Intravascular ultrasound to guide percutaneous interventions (Level of Evidence C)

### Class III:

- Routine preoperative coronary angiography (Level of Evidence C)
- Magnetic resonance imaging in haemodynamically unstable patients (Level of Evidence C)

## Recommendations for Diagnostic Imaging in Chronic Aortic Dissection



Class I:

- Magnetic resonance imaging (Level of Evidence C)
- Conventional angiography to guide percutaneous interventions (Level of Evidence C)

Class IIa:

- Transthoracic echocardiography followed by transoesophageal echocardiography (Level of Evidence C)
- Computed tomography (Level of Evidence C)
- Conventional angiography for pre-operative diagnosis in selected patients (Level of Evidence C)
- Conventional angiography for complete staging of the disease (Level of Evidence C)
- Intravascular ultrasound to guide percutaneous interventions (Level of Evidence C)

### Surgical and Interventional Therapy

#### Surgery in Acute Type A (Type I and II) Aortic Dissection

The aim of surgery is to prevent aortic rupture, pericardial tamponade, and to relieve aortic regurgitation. Implantation of a composite graft in the ascending aorta with or without reimplantation of coronary arteries is performed. A large variability of surgical procedures exist.

#### Recommendations for Surgical Therapy of Acute Type A (Type I and Type II) Aortic Dissection

Class I:

- Emergency surgery to avoid tamponade/aortic rupture (Level of Evidence C)
- Valve-preserving surgery: tubular graft if normal sized aortic root and no pathological changes of valve cusps (Level of Evidence C)
- Replacement of aorta and aortic valve (composite graft) if ectatic proximal aorta and/or pathological changes of valve/aortic wall (Level of Evidence C)

Class IIa

- Valve-sparing operations with aortic root remodeling for abnormal valves (Level of Evidence C)
- Valve preservation and aortic root remodeling in Marfan patients (Level of Evidence C)

#### Surgery in Acute Type B (Type III) Dissection

The indication for surgery in type B (type III) aortic dissection is limited to the following criteria:

- Persistent, recurrent chest pain

- Aortic expansion
- Periaortic haematoma
- Mediastinal haematoma

## Recommendations for Surgical Therapy of Acute Type B (Type III) Aortic Dissection

### Class I:

- Medical therapy (Level of Evidence C)
- Surgical aortic replacement if signs of persistent or recurrent pain, early expansion, peripheral ischaemic complications, rupture (Level of Evidence C)

### Class IIa:

- Surgical or endovascular fenestration and stenting if persisting mesenteric, renal or limb ischaemia or neurologic deficits (Level of Evidence C)

## Interventional Therapy in Aortic Dissection

- Interventional therapy in aortic dissection provides new approaches to handle complications.
- Aortic fenestration with or without stent placement allows immediate relief of organ malperfusion for:
  - Visceral
  - Renal
  - Limb ischaemia either before or after surgical treatment
- Graft stent implantation is an evolving technique which opens new avenues to treat type B (type III) dissection. Occlusion of entry tears induces thrombus formation and vessel wall healing.

## Recommendations for Interventional Therapy in Aortic Dissection

### Class IIa:

- Stenting of obstructed branch origin for static obstruction of branch artery (Level of Evidence C)
- Balloon fenestration of dissecting membrane plus stenting of aortic true lumen for dynamic obstruction (Level of Evidence C)
- Stenting to keep fenestration open (Level of Evidence C)
- Fenestration to provide re-entry tear for dead-end false lumen (Level of Evidence C)
- Stenting of true lumen to enlarge compressed true lumen (Level of Evidence C)

### Class IIb:

- Stenting of true lumen to seal entry (covered stent) (Level of Evidence C)

## Follow-up in Aortic Dissection

Close follow-up for aortic dissection by a specialized team includes the assessment of:

- Signs of aortic expansion
- Aneurysm formation
- Signs of leakages at anastomoses/stent sites
- Malperfusion

The single most important factor is excellent blood pressure control <135/80 mmHg. After hospital discharge regular outpatient visits at 1, 3, 6 and 12 month and thereafter every year are recommended. First choice is magnetic resonance imaging, second choice computed tomography and third choice transoesophageal echocardiography.

Recommendations for Prevention of Aortic Dissection in Inherited Diseases (Marfan's Syndrome, Ehlers-Danlos Syndrome, Annuloaortic Ectasia)

Class I:

- Life-long beta-adrenergic blockade (Level of Evidence C)
- Periodic routine imaging of the aorta (Level of Evidence C)
- Moderate restriction of physical activity (Level of Evidence C)

Class IIa:

- Prophylactic replacement of the aortic root before diameter exceeds 5.0 cm in patients with family history of dissection (Level of Evidence C)
- Prophylactic replacement of the aortic root before diameter exceeds 5.5 cm (Level of Evidence C)

Recommendations for Reoperation Following Repair of Aortic Dissection

Class I:

- Surgical intervention for:
  - Secondary aneurysm in dissected aorta remote from initial repair (Level of Evidence C)
  - Recurrent dissection or aneurysm formation at previous intervention site (Level of Evidence C)
- Graft replacement for gross dehiscence or infection (Level of Evidence C)

Class IIa:

- Use of homografts to replace infected prostheses (Level of Evidence C)
- Endovascular stenting if surgical indication and suitable anatomy (Level of Evidence C)

Recommendations for Therapy of Chronic Aortic Dissection

Class I:

Type A (Type I, II)

- Indications for surgery as in non-dissecting aneurysm if symptoms or aortic regurgitation or aortic diameter  $\geq 5.5$  cm (Level of Evidence C)

Type B (Type III):

- Indications for surgery as in non-dissecting aneurysms if symptoms or progressive aortic enlargement to  $\geq 6.0$  cm (Level of Evidence C)

Class IIa:

Type B (type III):

Endovascular stenting if surgical indication and suitable anatomy (Level of Evidence C)

#### Definitions:

Levels of Evidence:

- A. Data derived from multiple randomized clinical trials or meta-analyses
- B. Data derived from a single randomized trial or non-randomized studies
- C. Consensus opinion of the experts

Class of Recommendations:

Class I: Conditions for which there is evidence or general agreement that a given procedure or treatment is useful and effective

Class II: Conditions for which there is conflicting evidence or a divergence of opinion about the usefulness/efficacy of a procedure or treatment

Class IIa: Weight of evidence/opinion is in favour of usefulness/efficacy

Class IIb: Usefulness/efficacy is less well established by evidence/opinion

Class III: Conditions for which there is evidence and/or general agreement that the procedure/treatment is not useful/effective and in some cases may be harmful

CLINICAL ALGORITHM(S)

None provided

#### EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The level of evidence and class of recommendation is given for selected recommendations (see "Major Recommendations" field).

For diagnostic tests and surgical as well as interventional procedures related aortic dissection, no evidence-based medical data for group A and B were available, so that a grading concerning consensus between the members was chosen.

## BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

### POTENTIAL BENEFITS

- In general, this guideline may lead to better and earlier diagnosis of aortic diseases even in emergency situations, which could decrease cardiovascular mortality and morbidity.
- Surgical therapy may prevent aortic rupture or the development of pericardial effusion. Surgery may also eliminate aortic regurgitation, avoid myocardial ischemia, and relieve intractable pain.
- Using interventional therapy, flow could be restored in more than 90% of vessels obstructed from aortic dissection in one study.

### POTENTIAL HARMS

#### Vasodilators

- Vasodilators can increase the force of left ventricular ejection

#### Pericardiocentesis

- Pericardiocentesis as an initial therapeutic step before surgery may be harmful as this reduces intrapericardial pressure and may therefore cause recurrent bleeding

#### Aortography

- Aortography is invasive and therefore has an inherent risk. In addition, this technique requires the administration of potentially nephrotoxic radiopaque contrast media and ionizing radiation.
- In critically ill patients the time required for assembling the angiographic team, bringing the patient to the angiographic suite and the duration of the procedure itself may be too long to be justified. This should always be taken into account as diagnostic delays are associated with an increased mortality.

#### Surgical Therapy

- Implantation of allografts and xenografts should be restricted to elderly patients or special other indications since late postoperative degeneration may require reoperation on the aortic root
- Surgery carries the inherent risks of complications and mortality

#### Interventional Therapy (Percutaneous Balloon Fenestration and Stenting)

- Potential problems arise from unpredictable haemodynamic alterations in the true and false lumen after fenestration and stenting. These alterations may result in loss of previously well perfused arteries, as well as in loss of the desired salvage of compromised arteries.
- Paraplegia seems to occur, related to extensive graft stenting but not with short (<15 cm) stents and in consecutive instead of single approaches if longer segments have to be stented.
- Patients can develop an inflammatory reaction after implantation. This may present as an elevated C-reactive protein in combination with fever. Both signs may disappear spontaneously as the healing progresses.

## IMPLEMENTATION OF THE GUIDELINE

### DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

## INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

### IOM CARE NEED

Getting Better  
Living with Illness

### IOM DOMAIN

Effectiveness

## IDENTIFYING INFORMATION AND AVAILABILITY

### BIBLIOGRAPHIC SOURCE(S)

Erbel R, Alfonso F, Boileau C, Dirsch O, Eber B, Haverich A, Rakowski H, Struyven J, Radegran K, Sechtem U, Taylor J, Zollikofer C, Klein WW, Mulder B, Providencia LA. Diagnosis and management of aortic dissection. Eur Heart J 2001 Sep;22(18):1642-81. [295 references]

### ADAPTATION

Not applicable: The guideline was not adapted from another source.

### DATE RELEASED

2001 Sep

### GUIDELINE DEVELOPER(S)

European Society of Cardiology - Medical Specialty Society

## SOURCE(S) OF FUNDING

European Society of Cardiology

## GUIDELINE COMMITTEE

Task Force on Aortic Dissection

## COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Task Force members: R. Erbel (Chairman); F. Alfonso; C. Boileau; O. Dirsch; B. Eber; A. Haverich; H. Rakowski; J. Struyven; K. Radegran; U. Sechtem; J. Taylor; Ch. Zollikofer

## FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

## GUIDELINE STATUS

This is the current release of the guideline.

An update is not in progress at this time.

## GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the [European Society of Cardiology \(ESC\) Web site](#).

Print copies: Available from Elsevier Publishers Ltd. 32 Jamestown Road, London, NW1 7BY, United Kingdom. Tel +44.207.424.4200/ Tel: +44 207 424 4389; Fax: +44 207 424 4433; e-mail: [gr.davies@elsevier.com](mailto:gr.davies@elsevier.com); Web site: [www.escardiocontent.org](http://www.escardiocontent.org).

## AVAILABILITY OF COMPANION DOCUMENTS

The following is available:

- Recommendations for Task Force creation and report production. Sophia Antipolis (France): European Society of Cardiology, 2002.

Electronic copies: Available in Portable Document Format (PDF) from the [European Society of Cardiology \(ESC\) Web site](#).

## PATIENT RESOURCES

None available

## NGC STATUS

This summary was completed by ECRI on April 2, 2002.

#### COPYRIGHT STATEMENT

This summary is based on the original guideline, which is subject to the guideline developer's restrictions.

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The logo for FIRSTGOV, with "FIRST" in blue and "GOV" in red, and a small red star above the "I".

